Letters

Conjugate deviation of gaze in hepatic encephalopathy

Sir: Transient ocular motor disturbances such as ocular bobbing,¹ dysconjugate gaze,² skew deviation³ and absent horizontal responsiveness to oculovestibular testing⁴ have been described in patients with hepatic encephalopathy. Sustained conjugate deviation of the eyes, however, is usually considered indicative of a contralateral irritative lesion or an ipsilateral destructive lesion of the cortex or a lesion of the contralateral pontine gaze centre.⁵ Here we describe a patient with fulminating hepatic failure in whom conjugate deviation of gaze developed two days before death. No structural lesion could be demonstrated on CT scan or at necropsy.

A 52-year-old man with a long history of ethanol abuse and biopsy-proved hepatic cirrhosis was admitted in an obtunded state. On examination he had tense ascites and pedal oedema and heavily jaundiced sclerae. He responded to only simple verbal commands and had bilateral asterixis; neurological assessment was otherwise unremarkable. Laboratory data at that time were: total bilirubin 273mmol/l; serum albumin 16g/l; serum aspartate transaminase 518u/l, lactic dehydrogenase 452u/l; alkaline phosphatase 915u/l and γ -glutamyltransferase 430u/l; haemoglobin 9.3g/dl. Treatment with low protein diet, lactulose, and spironolactone was instituted. The patient's mental state deteriorated progressively over the next week and he became unresponsive to verbal stimuli but continued to respond purposefully to pain. At this time it was noticed that his eyes were deviated to the left in a conjugate fashion and failed to respond to caloric and oculovestibular stimuli. Pupils were equal and reactive to light and there was no papilloedema. Plantar responses were flexor and tone of the limbs was normal. Total bilirubin was now 316 μ mol/l and serum ammonia 116 μ mol/l (normal 17-47 μ mol/l).

A structural cortical or brainstem lesion was suspected. CT scan was normal and EEG showed nonspecific slowing in the theta range. CSF analysis showed only the presence of bilirubin in the fluid. The eyes remained deviated until two days later when the patient died suddenly. The last total bilirubin level a few hours before death was 522 μ mol/l.

A necropsy was performed. The brain (wet weight 1380 g) was jaundiced, but the most careful macroscopic and microscopic examination failed to reveal any structural lesion in any area. In keeping with the clinical picture, the liver was shrunken (840 g) and cirrhotic.

Stupor in this patient was due to hepatic encephalopathy. Cirrhosis with ascites proved by biopsy and necropsy, pedal oedema, abnormal liver function tests with elevated serum ammonia, diffuse EEG slowing and normal CT scan and CSF make this conclusion inescapable. The presence of sustained conjugate deviation of gaze, not previously described in hepatic stupor, was confusing and suggested a structural lesion. No such lesion could be demonstrated, either on CT scan or at necropsy. The occurrence of the conjugate deviation in this case must, presumably, be explicable on the basis of selective vulnerability to metabolic insult of centres associated with control of conjugate eye movements.

There is no doubt that toxic-metabolic conditions may affect brain-stem tegmental function as well as motor pathways and hemispheric structures.^{2,3} In such patients, systemic and laboratory evidence of severe metabolic disturbance and preservation of other functions at the same level should suggest a metabolic explanation for the signs.

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Recurrent meningitis due to labyrinthine fistula

Sir: We wish to describe two children who had recurrent meningitis. In children recurrent meningitis may have various causes.¹⁻³ In the absence of an immunological defect, it may be the result of anatomical abnormalities, such as skull fracture, tumour and congenital defects. In a few cases, the pathology responsible for the recurrent meningitis may be in the middle ear.⁴⁻⁸ The triad of recurrent meningitis, severe unilateral or bilateral sensori-neural deafness and opaque tympanic membrane or other evidence of fluid in the middle ear should raise the possibility of middle ear pathology. The two children we describe, both presented with this triad arising from labyrinthine fistulae.

A 5-year old girl with a normal birth history and development presented initially at 7 months of age with *Haemophilus influenzae* meningitis. She recovered quickly but developed severe bilateral sensori-neural deafness. A few weeks later, she had an attack of pneumococcal meningitis which recurred at the age of 23 months. Radiological examination revealed a defect at the apex of the petrous temporal bone. A left lateral craniotomy with dural repair to the middle fossa was performed at 2 years of age. However, she had five further attacks of meningitis which included three due to pneumococcus and two due to *Haemophilus influenzae* at between 2 years 3 months and 5 years 6 months. At the age of 5 years 8 months she was admitted to the Hospital for Sick Children, London. Examination showed the left tympanic membrane to be normal, but the right was bluish and a fluid level was seen. She had bilateral sensori-neural deafness. The rest of the examination, including full neurological examination, was normal. The results of investigations were not diagnostic: haemoglobin level was 11.9g/dl; white cell count was $18.7 \times 10^9/l$ (neutrophils 71%; lymphocytes 21%; reticulocytes 5%; eosinophils 2%; basophils 1%). Complement C3 was 108% of standard. Nitroblue tetrazolium test was normal. Radiographs of the sinuses showed minor mucosal thickening in the antrum. Chest radiograph was normal. Tomograms of the mastoids showed the lateral part of the right internal auditory meatus wider than that on the left, and the vestibule was slightly larger. The basal coil of the right cochlea was normal. Computed tomography (CT) showed an opaque right middle ear. A right exploratory

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tympanotomy was performed and a fistula was found in the foot plate of the stapes. The stapes was removed and the oval window was plugged with temporal fascia. Since then (18 months) no further attacks of meningitis have occurred.

The second case was a girl, born following a normal pregnancy, and delivery, to healthy unrelated parents. Her development was normal except for deafness discovered at 12 months of age. She was fitted with a hearing aid in both ears. She has had three attacks of pneumococcal meningitis at the age of 21 months, 23 months, and 27 months, and was referred to the Hospital for Sick Children. Examination revealed a normal healthy child with a hearing aid in both ears. Throat, nose and right ear were normal; the left ear showed air bubbles and fluid in the middle ear. The tympanic membrane was dull. There was no other abnormality. Her investigations also were not diagnostic: haemoglobin and white cell count were normal. Serum immunoglobulins IgG and IgA were normal, IgM slightly raised. Nitroblue tetrazolium test was normal; cerebrospinal fluid (CSF) showed no cells and no organisms were cultured. Protein was 0.1 g/l (normal); skull radiographs and tomograms of the mastoids were normal. Metrizamide cisternogram did not demonstrate a leak into sinuses, and was normal. CSF flowed through the subarachnoid space. A technetium brain scan also showed normal CSF distribution and no abnormal accumulation was demonstrated. Hearing test confirmed severe bilateral sensorineural hearing loss. She also had an exploratory tympanotomy, and this showed a leak of perilymph from the central part of the footplate of the stapes. The stapes was removed from the oval window and the cavity was plugged with tragal perichondrium and post-auricular muscle. She has remained free from attacks of meningitis for the past 42 months.

Recurrent meningitis in the presence of other systemic infections may indicate an immunological disorder. However, when it occurs without systemic infections, an anatomical defect is likely. When confronted with a child with the triad recurrent meningitis, hearing loss (especially sensori-neural) and evidence of middle ear pathology even without abnormality in the tomographic study of temporal bones, then labyrinthine fistula should be suspected. Detailed investigations, as in both our cases, may fail to show a site of CSF leak. Therefore in such a child exploratory tympanotomy should be undertaken.

We thank Mr John Evans and Mr Robert

Pracy for allowing us to report these children.

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Errata

In the October issue of the *Journal of Neurology, Neurosurgery and Psychiatry* the letters by Quinn *et al* and by Plant were printed without their figures. We apologise to these authors and publish below the letters in full.

Insulin-induced hypoglycaemia does not abolish chorea

Sir: Pathological changes occur in the hypothalamus in Huntington's disease.¹ Insulin tolerance tests have been used to examine hypothalamic function in such patients, and mild abnormalities of growth hormone secretion have been described.²⁻⁴ In the course of such an investigation, Keogh *et al*² noted that chorea ceased some 30 min after the insulin injection and was not evident for the next 60 to 75 min in all of the twelve patients studied. They did not think that this dramatic change was due to an altered level of consciousness, for "all patients were awake throughout the investigations and were checked repeatedly to see that they were capable of verbal communication". Subsequently, Lavin *et al*³ described similar observations in another group of eight patients with Huntington's disease, in all of whom chorea disappeared for at least an hour within about half-an-

hour of the insulin injection. Such a dramatic effect on chorea might provide some clue as to the pathophysiology of that movement disorder, so we have repeated the study concentrating on the effect of insulin-induced hypoglycaemia on the chorea.

Five patients with Huntington's disease (four males and one female; aged 30 to 70 years; with disease duration from 2 to 13 years; four on no drugs and one on tetrabenazine 25 mg three times daily) with obvious chorea were studied. After an overnight fast, blood was withdrawn for glucose estimation, and insulin (0.1 mg/kg) was injected into the opposite arm. Blood sugar and clinical response were measured every 10 min for 60 min, and then every 20 min for a further 60 min. The severity of chorea was rated using a specially designed scale described in detail elsewhere.⁵ In addition the number of choreic movements occurring at rest in one selected region, such as an eye, finger or toe depending on the individual patient, was counted over a 60 sec period. Blood sugar fell below 2.0 mmol/l and symptoms and/or signs of hypoglycaemia developed in all subjects. However, the intensity of chorea did not alter. Three subjects fell asleep during the test, and chorea disappeared in two, but on arousal their chorea was of the same severity as before insulin. Unfortunately, insulin-induced hypoglycaemia had no effect on chorea in our patients.

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